

Atrial myxoma as a cause of stroke: case report and discussion

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Abstract

Cardiac myxoma is a source of emboli to the central nervous system and elsewhere in the vascular tree. However, nonspecific systemic symptoms and minor embolic phenomena may be overlooked in the absence of any history of cardiac problems. In this situation, cardiac investigations may not be performed, and diagnosis of this rare condition may be delayed until the onset of more significant embolic disease, such as stroke with functional impairment, as in the case reported here. The clinical presentation of cardiac myxoma is discussed, along with appropriate investigations and treatment, which may prevent such sequelae.

Case

A 48-year-old man who presented to the emergency department reported a fall and transient loss of consciousness. The physical examination was limited by the patient's confusional state, but it revealed weakness of the right arm and leg. No seizure had been witnessed. He smoked but had no other conventional vascular risk factors such as hypertension, diabetes or dyslipidemia.

Over the previous year he had reported 4 or 5 syncopal episodes, weight loss of 20 lb (about 9 kg) to 130 lb (about 59 kg), and symptoms of myalgia and arthralgia. His family reported gradual memory impairment and personality change. Investigations before the current event had focused on a possible rheumatological disorder and had revealed elevation of the erythrocyte sedimentation rate (ESR; peak 74 mm/h), a positive speckled pattern on antinuclear antibody (ANA) testing and a weakly positive result on classical antineutrophil cytoplasmic antibody (c-ANCA) testing. The results of biopsy of a lower extremity rash were suggestive of livedo reticularis. An autoimmune process such as Wegener's granulomatosis or systemic lupus erythematosus had been suggested, and the patient had started steroid and chloroquine therapy but without improvement.

During the current admission, repeat ANA and c-ANCA testing yielded negative results. The ESR remained elevated at 64 mm/h. Electrocardiography (ECG) demonstrated sinus rhythm with left anterior fascicular block. CT of the brain showed previous infarcts of the right parietal and left occipital lobes. MRI demonstrated additional old bihemispheric infarcts and a more recent infarct in the left frontal lobe on apparent diffusion coefficient images (Fig. 1); the latter was the most likely source of the patient's acute symptoms. The results of magnetic resonance angiography (MRA) were unremarkable.

Cerebral angiography revealed multiple, bilateral fusiform aneurysms throughout the anterior and posterior circulations (Fig. 2). These findings, along with the bilateral distribution of cerebral infarction, suggested a proximal embolic source. Transthoracic echocardiography iden-

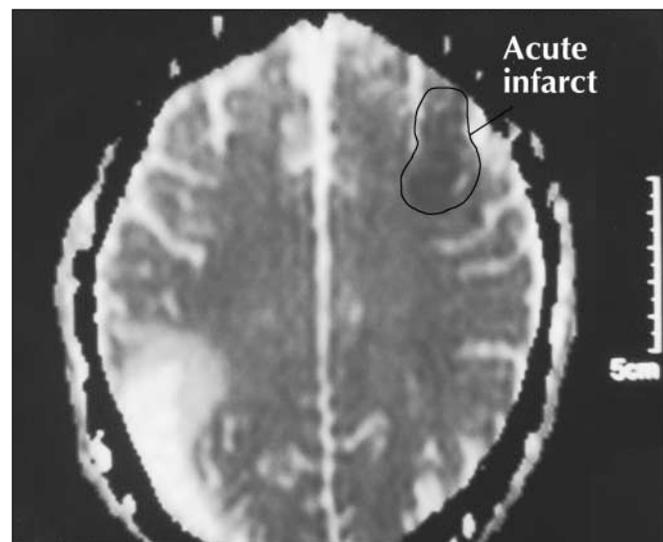


Fig. 1: MRI of the brain (apparent diffusion coefficient image) shows old bihemispheric infarcts and a more recent left frontal infarct (highlighted).

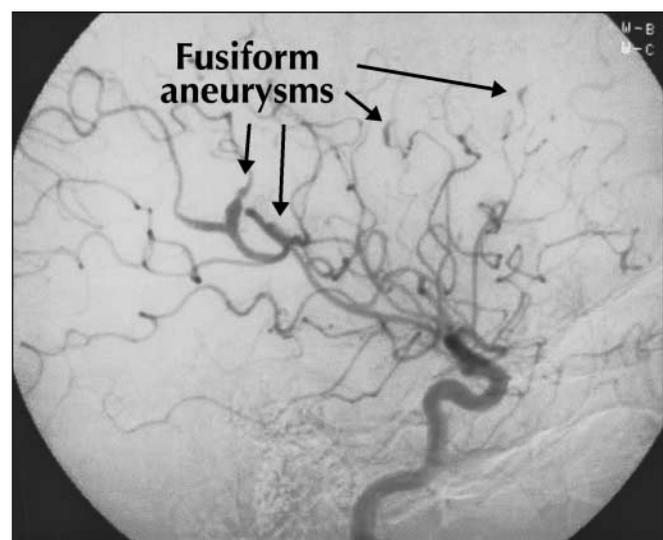


Fig. 2: Cerebral angiogram shows multiple fusiform aneurysms (highlighted).

tified a left atrial myxoma (4 × 2.5 × 2.5 cm) prolapsing through the mitral valve in diastole (Fig. 3).

The myxoma, of which there was no known family history, was resected without complications 11 days after admission. Steroids were gradually reduced and stopped postoperatively. The ESR fell initially to 16 mm/h and then to 4 mm/h at follow-up 3 months after the surgery. The patient was subsequently referred for rehabilitation for residual right-sided weakness and gait unsteadiness. Formal neuropsychological testing demonstrated deficits in concentration, attention, executive function, visuospatial function and memory. These were thought to be consistent with the ischemic brain damage.

Comments

Atrial myxoma, the most common benign cardiac tumour, is found more commonly in young adults with stroke or transient ischemic attack (1 in 250) than in older patients with these problems (1 in 750).¹ The annual incidence is 0.5 per million population,² with 75% of cases occurring in the left atrium. There is a 2:1 female preponderance,³ and the age at onset is usually between 30 and 60 years. Delay in diagnosis from symptom onset may range from 1 to 126 months.³

Although atrial myxoma is mostly sporadic, at least 7% of cases are familial.⁴ The best described familial type is Carney complex, characterized by cutaneous spotty pigmentation, cutaneous and cardiac myxomas, nonmyxomatous extracardiac tumours and endocrinopathies. It is transmitted in an autosomal dominant manner, through a causative mutation of the *PRKAR1α* gene located on the long arm of chromosome 17 (17q22-24 region).⁵

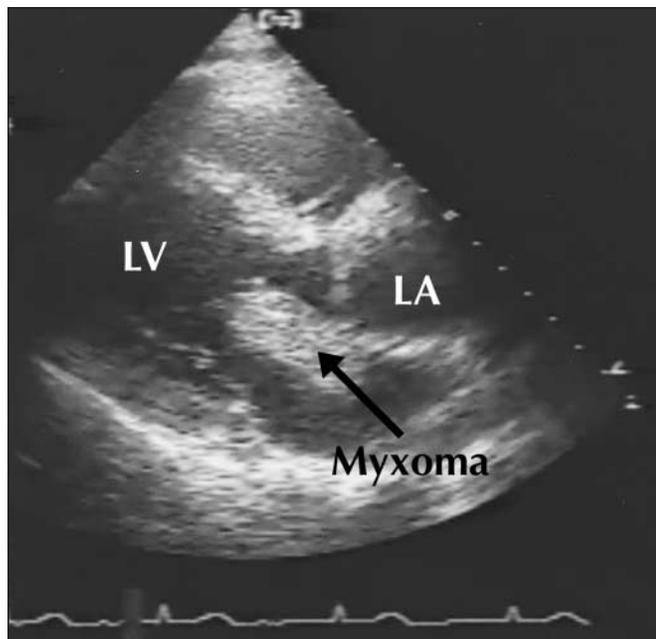


Fig. 3: Echocardiogram shows the left atrial myxoma (highlighted). LV = left ventricle, LA = left atrium.

The presentation of atrial myxoma often comprises a diagnostic triad (Table 1). Active illness is often accompanied by elevation of ESR and C-reactive protein, hyperglobulinemia and anemia. Constitutional symptoms may be mediated by interleukin-6, produced by the myxoma itself.⁹

Strokes are often recurrent¹⁰ and may be embolic or hemorrhagic, the presentation ranging from progressive multi-infarct dementia¹¹ to massive embolic stroke causing death.¹² Because tumour fragments or adherent thrombus may embolize, anticoagulation may not be protective.¹³ The multiple, bilateral fusiform aneurysms¹⁴ commonly found on peripheral arterial branches predispose the patient to cerebral hemorrhage.¹⁴ As in the patient described here, MRA may miss intracranial aneurysms less than 3 mm in diameter and is inferior to conventional angiography in this regard.¹⁵

The presence of embolic phenomena, especially in young patients with neurological symptoms, should prompt early neuroimaging and echocardiography, even in the absence of electrocardiographic or auscultation abnormalities.

The presence of embolic phenomena, especially in young patients with neurological symptoms, should prompt early neuroimaging and echocardiography, even in the absence of ECG or auscultation abnormalities. Auscultation abnormalities may be absent in 36% of patients with myxoma, and a murmur suggestive of mitral stenosis has been reported in only 54%.¹ Transesophageal echocardiography, which has been reported as having 100% sensitivity for cardiac myxoma,¹⁶ is preferred over transthoracic echocardiography. Transesophageal echocardiography may also improve the detection of other major cardioembolic sources (e.g., intracardiac thrombus, vegetations or aortic arch plaque), as well as less common potential sources (e.g., patent foramen ovale, atrial septal aneurysm or left ventricular aneurysm).¹⁷

Cardiac MRI can assist in delineating tumour size, attachment and mobility.¹⁸ This information may be helpful in surgical resection, which, because of the risk of further embolization, should not be deferred even in asymptomatic cases discovered incidentally. Resection may lead to normalization of serum interleukin-6 levels and resolution of constitutional symptoms,⁹ and the intracranial aneurysms may regress and resolve.¹⁹

Neurological sequelae after resection are rare but may occur without recurrence of the cardiac tumour. Instead of regressing, aneurysms may enlarge or appear for the first time.²⁰ Tumour fragments that have metastasized to the

Table 1: Diagnostic triad in presentation of atrial myxoma

Feature	Manifestations	Frequency, % of patients
Obstructive symptoms ^{6,7}	Heart failure, dyspnea, syncope, sudden death (rare)	54–95
Constitutional symptoms ^{3,6}	May mimic autoimmune disease or vasculitis: myalgia, arthralgia, weight loss, fatigue, fever, Raynaud's phenomenon, finger clubbing	34–90
Embolic phenomena ⁶⁻⁸	Emboli may travel to any organ, but 73% reach central nervous system, including spinal cord	10–45

vessel walls may enlarge, causing vessel occlusion and delayed infarction,¹⁴ or they may penetrate through the vessel wall, forming intra-axial metastases.²¹

Primary tumours recur in only 1% to 3% of sporadic cases, often because of inadequate resection.⁴ For patients with sporadic myxoma, annual review with echocardiography is suggested for a period of 3 to 4 years, when the risk of recurrence is greatest.²² For Carney complex, which has a recurrence rate of up to 25%, lifetime annual review with familial screening is recommended.²³

Conclusions

The diagnosis of atrial myxoma can be elusive, especially when the symptoms suggest a systemic illness. In the case reported here, the significance of these symptoms became apparent when the patient presented acutely with a motor deficit as a result of cerebrovascular embolism. The presence of bihemispheric infarction focused subsequent investigations on the possibility of a proximal source of embolization, which resulted in identification of the causative atrial myxoma.

This article has been peer reviewed.

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Competing interests: None declared.

Contributors: Drs. O'Rourke, Dean and Mouradian were responsible for the conception of the article, the literature search and writing of the initial drafts. Drs. Akhtar and Shuaib reviewed and contributed suggestions and revisions to the final drafts of the article.

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